

Idiopathic retroperitoneal fibrosis - A rare cause of biliary obstruction

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Section: Abdominal imaging

Area of Interest: Abdomen

Procedure: Diagnostic procedure

Imaging Technique: MR

Imaging Technique: CT-Angiography

Imaging Technique: CT

Special Focus: Acute Case Type: Clinical Cases

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Patient: 58 years, male

Clinical History:

A 58-year-old man presented with itching, weakness, jaundice and weight loss. On clinical examination, abdomen was soft, non-tender with a vague lump in the right hypochondriac region. No history of fever, arthralgia or any drug ingestion like methysergide and ergotamine was noted.

Imaging Findings:

Ultrasonography revealed dilated intrahepatic biliary radicles and bilateral hydroureteronephrosis. Subsequently, MRCP was done, which revealed moderate dilatation of bi-lobar intrahepatic duct with involvement of CHD along with bilateral hydroureteronephrosis. (Fig. 1, 2) Based on these findings, D-J stenting was done. Abdominal CT angiography revealed thickened enhancing CBD wall with stricture of CHD confluence causing dilatation of intrahepatic biliary radicles and an enhancing periaortic soft tissue mass involving SMA, left renal vein, bilateral iliac vessels and encasing bilateral ureters. (Fig 3a-d, 4) Thereafter, ERCP-guided papillotomy was done and CBD stent was placed.

Elective laparotomy with Roux-en-y- hepatico-jejunostomy was done. Histopathological correlation of the resected CBD was done which revealed extensive fibrosis with presence of collagenisation admixed with numerous inflammatory cells. (Fig. 5) IgG4 assay was not performed. Follow-up CT scan done after the course of steroid showed significant regression of soft tissue around the aorta and iliac arteries. (Fig 6a, 6b)

Discussion:

Idiopathic retroperitoneal fibrosis (IRF), also known as Ormond's disease, is an extremely unusual disorder with an incidence of 1.3:1000000. The usual age of diagnosis is around 40 to 60 years with men being affected two to three times more often than women. [1] It is characterised by deposition of peri-aortic fibrous tissue involving the retroperitoneal structures including ureters. Occasionally it may involve the mesentery, small bowel, pancreas and more rarely the biliary system. [1]

The aetiology of retroperitoneal fibrosis (RPF) is unknown in about two-thirds of cases. The probable pathophysiology is an autoimmune disorder associated with antibodies against ceroid, a complex of oxidised lipid and protein. [2] Ormond's disease when associated with sclerosing cholangitis is also termed IgG4 associated cholangitis (IAC) due to presence of high levels of IgG and IgG4. [3] It has an insidious clinical onset with

nonspecific complaints of abdominal pain not responding to anti-inflammatory drugs.

RPF appears diffusely hypoechoic periaortic lesion on ultrasound. On CT, RPF appears as a diffuse fibrous plaque, isodense with striated muscle showing variable enhancement on contrast administration depending on the stage of fibrotic process. The degree of enhancement decreases with the chronicity of disease giving a rough estimate of the stage of the disease process. MRI is slightly superior to CT in defining the lesion as it provides better contrast resolution. Retroperitoneal fibrosis appears as homogeneously low signal intensity on T1-weighted imaging. However, the T2 signal may vary depending upon the degree of active inflammation. Thus chronic inactive fibrosis will have low signal on both T1- and T2-weighted imaging owing to a lesser degree of oedema. [4, 5]

Till now, only 15 cases have been reported in which biliary tract has been involved in IRF. [6] In this case, the patient presented with symptoms of cholestasis secondary to involvement of the extrahepatic biliary tree. Stricture of the common bile duct was noted along with pain in the abdomen and jaundice mimicking features of cholangiocarcinoma. Additionally, on ERCP presence of multiple strictures on intra and extrahepatic biliary tree can resemble the appearance of primary sclerosing cholangitis (PSC). It is therefore crucial to discriminate IRF from PSC as their treatment options are different. IRF shows a good response to corticosteroids unlike PSC. The present case highlights the rare involvement of the biliary tract in retroperitoneal fibrosis with the patient presenting with symptoms of cholestasis. Familiarity with the range of findings in different imaging modalities is critical for timely counselling, appropriate management and effective treatment.

Differential Diagnosis List: Retroperitoneal fibrosis involving SMA, renal vein, ureters and bile ducts., Cholangiocarcinoma, Primary sclerosing cholangitis

Final Diagnosis: Retroperitoneal fibrosis involving SMA, renal vein, ureters and bile ducts.

References:

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Figure 1

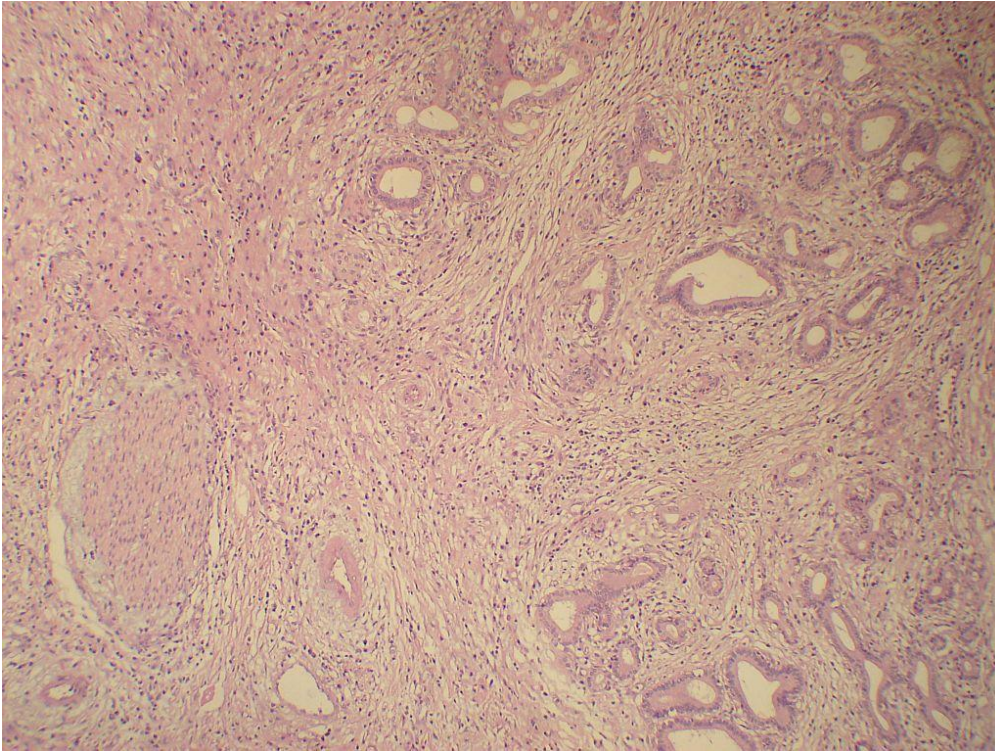
a



Description: Sagittal oblique reconstructed CT image showing diffuse circumferential soft tissue involving common bile duct and causing its narrowing. **Origin:** Department of Radiology, BLK Superspeciality Hospital, New Delhi, India

Figure 2

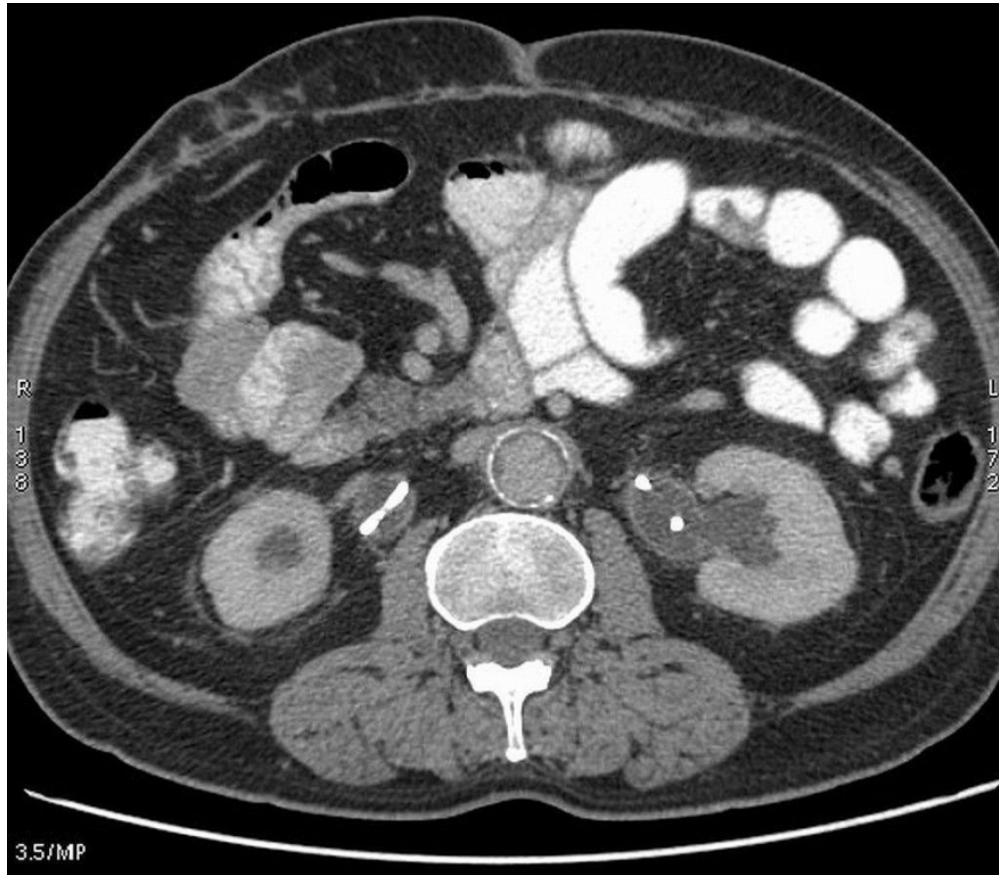
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Description: Microscopic image showing thickening of common bile duct wall with extensive fibrosis, area of collagenisation admixed with lymphoplasmocytes infiltrates. **Origin:** Department of Radiology, BLK Superspeciality Hospital, New Delhi, India

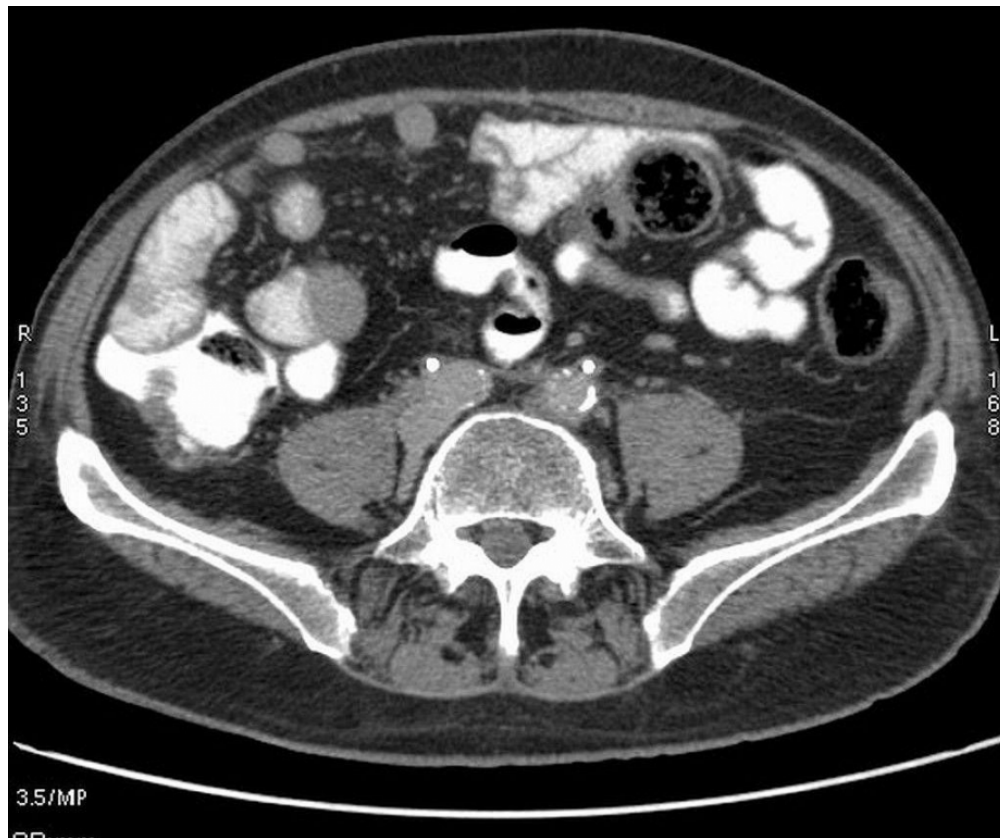
Figure 3

a



Description: Contrast-enhanced axial section CT showing decrease in peri-aortic soft tissue after steroid treatment. **Origin:** Department of Radiology, BLK Superspeciality Hospital, New Delhi, India

b



Description: Contrast-enhanced axial section CT showing decrease in soft tissue around bilateral common iliac arteries after steroid treatment. **Origin:** Department of Radiology, BLK Superspeciality Hospital, New Delhi, India

Figure 4

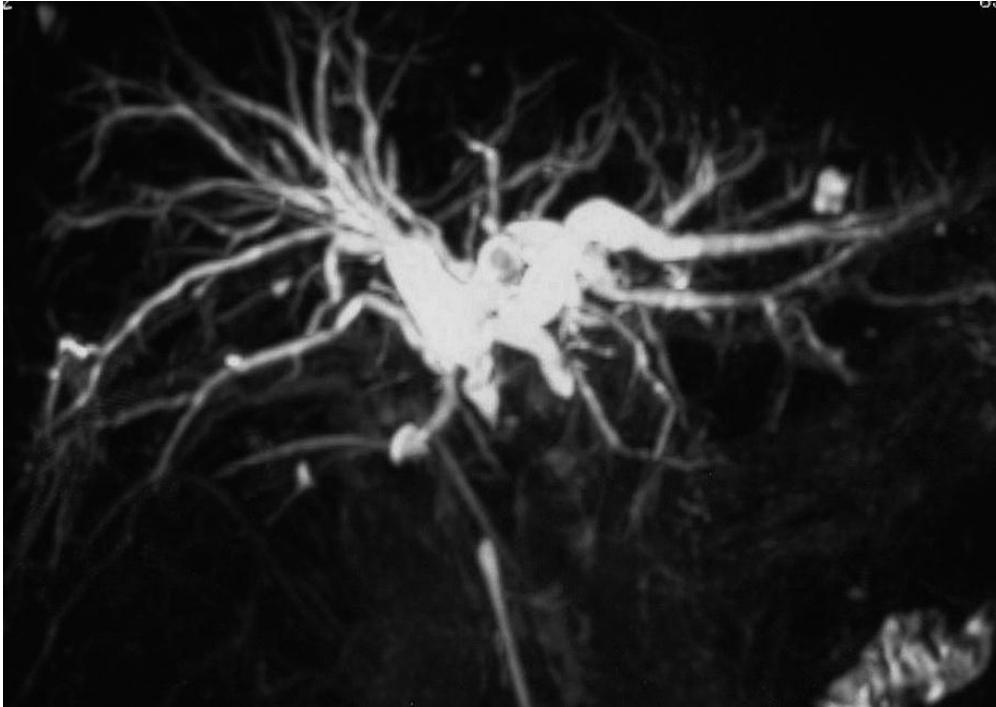
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Description: Axial T2-weighted MR showing bilateral hydronephrosis. **Origin:** Department of Radiology, BLK Superspeciality Hospital, New Delhi, India

Figure 5

a

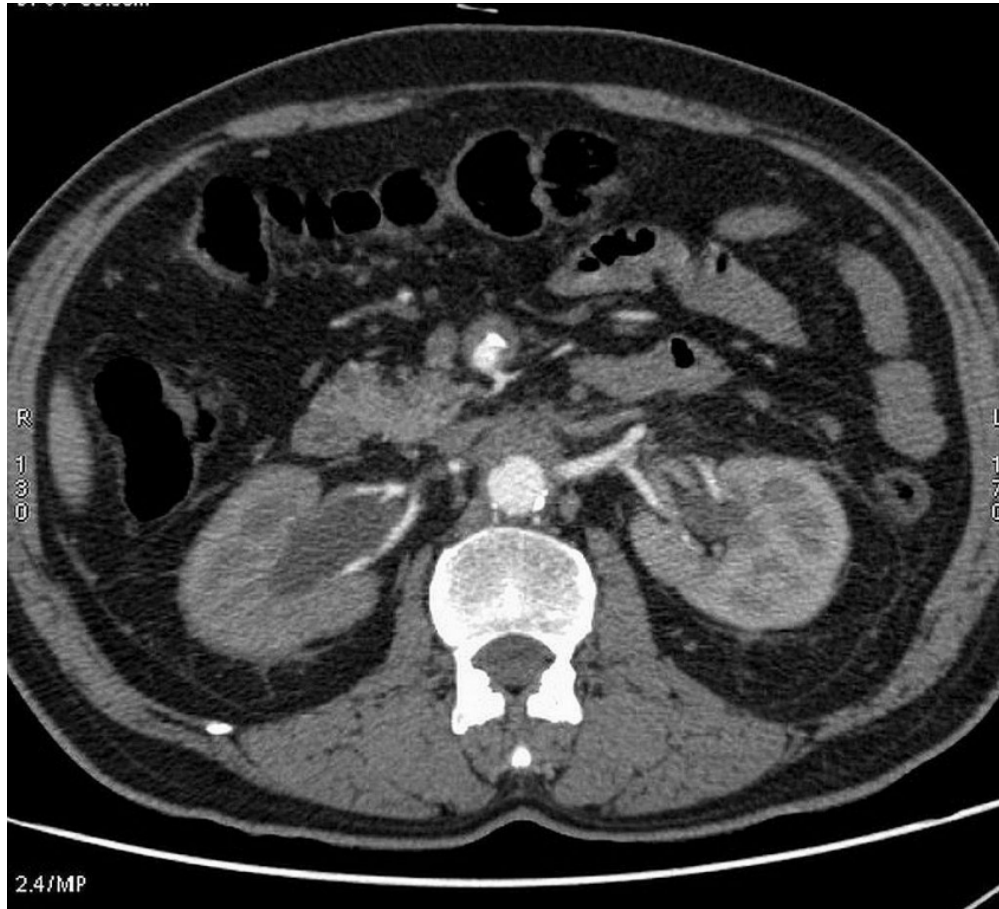


Description: 3D MRCP depicting gross dilatation of biliary radicles caused by narrowing at CBD.

Origin: Department of Radiology, BLK Superspeciality Hospital, New Delhi, India

Figure 6

a



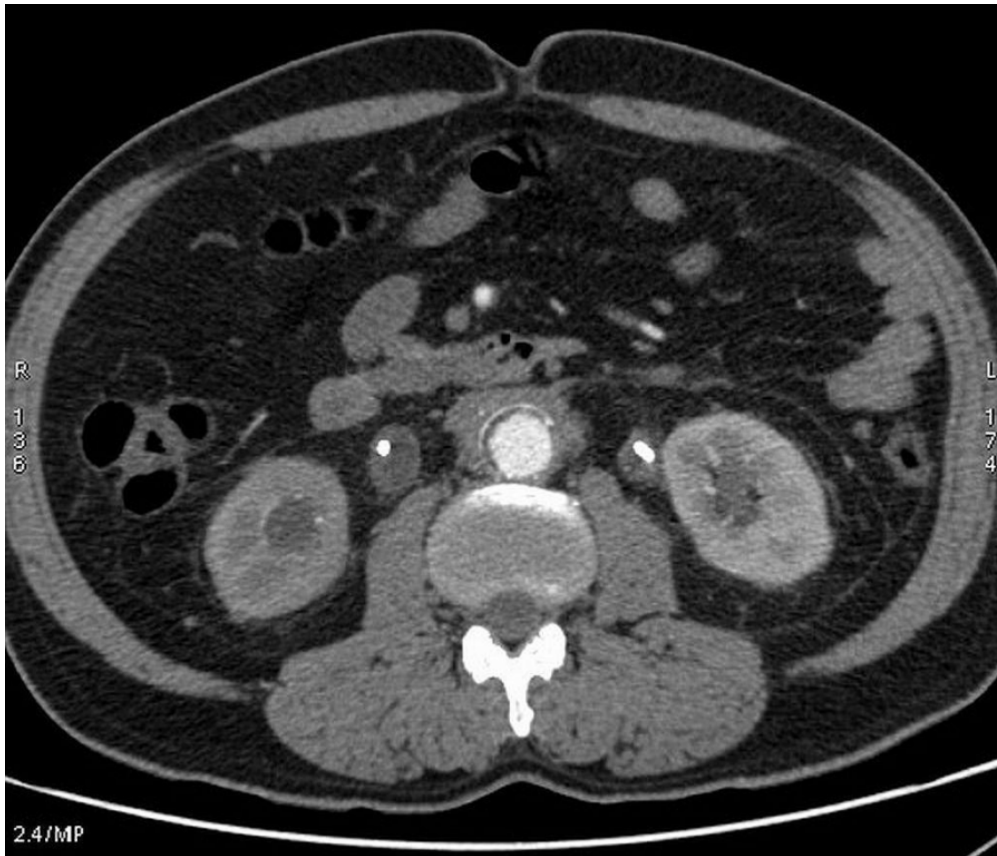
Description: Contrast-enhanced axial sections showing enhancing soft tissue around superior mesenteric artery. **Origin:** Department of Radiology, BLK Superspeciality Hospital, New Delhi, India

b



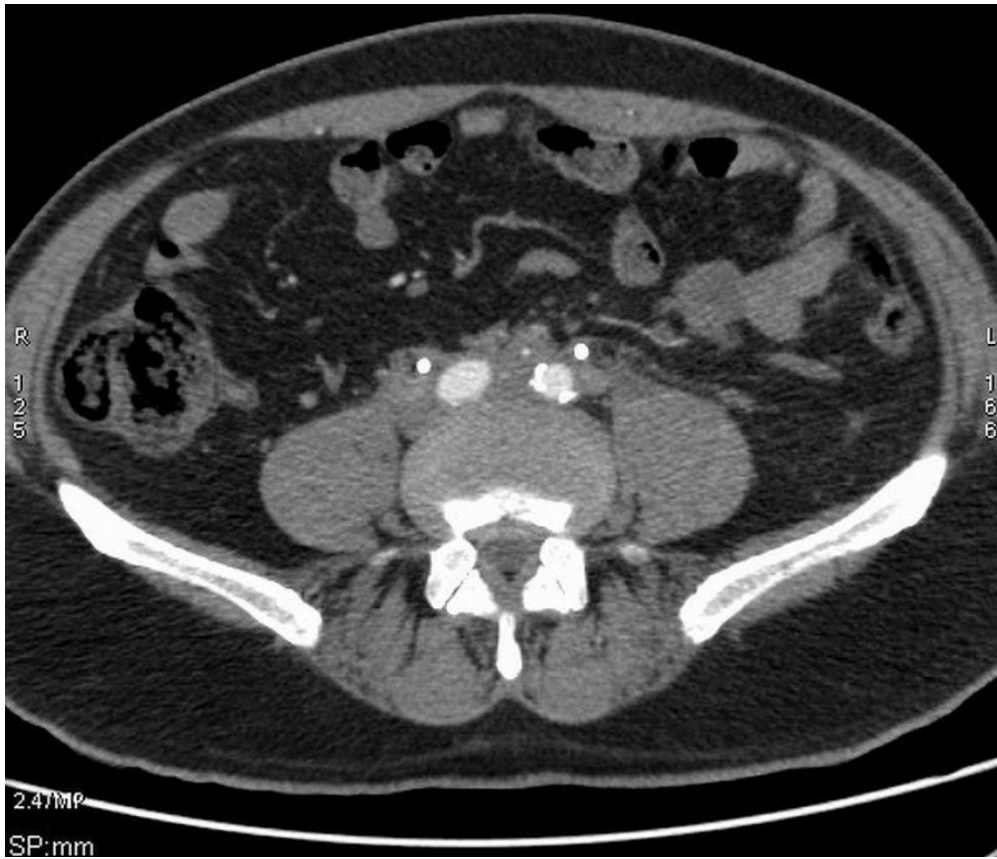
Description: Contrast-enhanced axial section CT showing enhancing soft tissue encasing and narrowing left renal vein. Bilateral DJ stent in situ. **Origin:** Department of Radiology, BLK Superspeciality Hospital, New Delhi, India

c



Description: Contrast-enhanced axial section CT showing abnormal enhancing peri-aortic soft tissue. Bilateral DJ stents are noted in situ. **Origin:** Department of Radiology, BLK Superspeciality Hospital, New Delhi, India

d



Description: Contrast-enhanced axial section CT showing abnormal enhancing soft tissue around the common iliac vessels. **Origin:** Department of Radiology, BLK Superspeciality Hospital, New Delhi, India